Spina Bifida

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Definition

Spina bifida is one of a group of birth defects known as **neural tube defects** (NTD). Within 28 days after conception, a tissue called the neural plate in the developing embryo "folds" and forms a tube. The tube then develops into the spinal cord. Spina bifida occurs when the neural plate folds incorrectly. This results in an abnormally formed section of the **spinal column** (backbone).

The affected section of the spinal column is easily recognized at birth. One or more **vertebrae** (bones of the spine) may be incompletely formed and the nerves (**spinal cord**) within the vertebrae protrude from the back. Thus the nerves of the spinal cord which carry messages from the brain to the rest of the body may be damaged. The point where the spinal cord protrudes from the back is called the **lesion**. The location on the vertebral column where the abnormality occurs is called the **lesion level**. For example, if the lesion occurred at the fourth lumbar vertebra then this is called a L-4 lesion.

Causes

The causes of spina bifida are complex and not well understood. However, most researchers agree that a number of factors are involved. These include one or more environmental factors that may coincide, in some persons, with an underlying genetic predisposition.

Prevalence and Incidence

The Spina Bifida Association of America estimates that there are more than 70,000 persons living in the United States who have this birth defect. In Arkansas, the Arkansas Spinal Cord Commission lists 485 persons with spina bifida on their service registry.

For the United States, spina bifida occurs in 7 out of every 10,000 live births. In Arkansas, the Center for Birth Defects Prevention (ACBDP) has calculated a rate of 5.1 per 10,000 live births for the years 1998-2000. For this time period, ACBDP indicates that 57 persons were born with spina bifida in Arkansas.

Risk Factors

Any woman who is of childbearing age is potentially at risk of having a pregnancy affected by spina bifida. In fact, 95 percent of all spina bifida pregnancies occur in women who have no history of this defect in their personal or family history.

However, some women are at higher risk to have a spina bifida affected pregnancy than others. Research has identified the following as risk factors:

- A previous pregnancy affected with spina bifida or another neural tube defect is about twenty times more likely to have another affected pregnancy than a woman without an affected pregnancy history.

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- Insulin-dependent diabetes
- Use of certain anti-seizure medication (valproic acid/Depakene and Carbamazapine/Tegretol)
- Medically diagnosed obesity
- High temperatures in early pregnancy (i.e., hot tube use and prolonged temperature)
- Race/ethnicity - NTDs are more common among white women than black women and are more common among Hispanic women than non-Hispanic women.
- Women of lower socioeconomic status.

Symptoms

A person with spina bifida will, to some extent, experience loss of bodily control below the level of lesion. The higher up (towards the head) the lesion is the greater the severity will be. Persons with spina bifida often experience:

- Loss of bowel control
- Loss of bladder control
- Loss of sensation
- Paralysis

In addition to the lesion, other parts of the central nervous system are likely to be abnormally formed. About 80 percent of persons with spina bifida have **hydrocephalus** or an excessive accumulation of cerebral spinal fluid in the head. The excessive fluid affects the formation of the brain and may lead to mental retardation unless surgery is undertaken to repair the abnormality and/or a shunt installed to drain the fluid. A **shunt** is a flexible plastic tube which drains the excessive fluid to another part of the body where it is absorbed and eliminated in the urine.

Arnold Chiari Malformation is another condition frequently experienced by persons with spina bifida. Part of the brain, the cerebellum, located in the back of the head extends down abnormally into the spinal canal. The cerebellum puts pressure on the spinal cord and/or blocks the flow of cerebral spinal fluid down the spinal cord. Surgery is necessary to correct the abnormality.

Types of Spina Bifida and Diagnosis

There are three types of spina bifida; two are readily diagnosed at birth but the third one is usually discovered later in life.

Spina bifida cystica (cyst-like) refers to the cyst or sac protruding from the back containing spinal tissues and fluid. The sac is easily noticed at birth and presents in two forms:

Meningocele is the least common. In this form, the sac contains fluid and tissues that cover the cord. The cord, itself is not in the sac; thus, it often is not damaged.

Myelomeningocele is the most common. Here, the sac contains not only fluid and covering tissues but also nerves of the spinal cord. Thus, the cord is likely to be damaged or not properly developed.

Spina bifida occulta (hidden) is not readily apparent at birth–no spinal column abnormality is visible. It rarely presents a problem for the estimated 5 to 10 percent of the general population who have it. The condition may not be discovered until much later in life when an x-ray reveals a thinning or a mild distortion of a part of the backbone.

Treatment

A newborn with spina bifida needs surgical repair of the lesion shortly after birth. In addition, separate surgeries may be required for installation of a shunt to control any hydrocephalus and/or to correct any abnormalities if Arnold Chiari Malformation is present.

A trunk/body brace may be necessary to encourage proper alignment of the spine and leg braces may be required to encourage proper development of the legs. As the child grows, larger braces will be required. Later, a wheelchair may be necessary for mobility. Also, glasses may be needed to correct

any eye/vision problems.

Around 15 to 30 percent of persons with spina bifida develop an allergy to latex acquired through latex exposure in multiple surgeries. Care should be taken to avoid contact with latex products if the allergy is present.

Most children attend regular school although special education classes may be needed for some subjects.

Prognosis

Persons with spina bifida can expect to live a normal life aided by assistive devices, such as, a wheelchair for mobility. Individuals often experience problems transitioning from adolescence to adulthood. Over protective parents can discourage the level of independency needed for adult life.

There is little data on the current life expectancy of persons with spina bifida. Before the introduction of shunts in the 1950's individuals rarely lived beyond a few years. Now, with the wide spread use of shunt technology, individuals can expect to live decades into adulthood. The life expectancy of persons with spina bifida lengthens every year as more and more recipients of shunt and other medical/surgical technology age.

Prevention

Research efforts around the world have discovered that folic acid, a common B vitamin, can prevent up to 70 percent of neural tube defects including spina bifida. In its natural form, folate, is found in orange juice, leafy green vegetables, beans and other foods.

In a public health effort, the United States Food and Drug Administration ordered all cereal grain products (wheat and corn flour) sold in the United States after October 1, 1998, to be fortified with folic acid (the synthetic form of folate). This action has increased the amount of folate consumed by the general population and reduced the occurrence of spina bifida by 31 percent (as compared to prefortification figures).

However, even with supplementation, many women in the United States do not get enough folate in their diet. That is why the Centers for Disease Control recommends that all women of childbearing age take a vitamin supplement containing 400mg of folic acid every day.

More Information

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